

case report

Primary CNS lymphoplasmacytic lymphoma: A case report and review of literature

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Lymphoplasmacytic lymphoma is a chronic lymphoproliferative disorder characterized by a proliferation of plasma cells, small lymphocytes, plasmacytoid lymphocytes and the production of monoclonal IgM. Primary central nervous system lymphomas (PCNSL) are rare non-Hodgkin lymphomas (NHL) that can be found in the brain, leptomeninges, eyes or spinal cord, and are mostly intracerebral. PCNSLs constitute 3–4% of primary brain tumors, and in most cases are diffuse large B-cell lymphomas (DLBCL).¹ Low grade lymphomas as primary central nervous system (CNS) lymphoma are very rare. We present here a case report of a woman who presented with headache and was found to have primary intracranial lymphoplasmacytic lymphoma (LPL).

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CASE REPORT

A 53-year-old woman presented to emergency department with a 3–4 month history of worsening headaches on left side associated with occasional blurriness of vision and tenderness to touch on left side of her scalp. She denied photophobia, stiffness

of neck, fever, nausea or vomiting; there was no variation in symptoms by position or movement. One year prior, she had a cervical discectomy and fusion of C3–C7. Her past medical history included high blood pressure and dyslipidemia. She was a former smoker; her family history was non-contributory. Physical exam did not show any focal neurologic signs, and strength in all the extremities was normal. Non-contrast CT scan of head done in the emergency department was normal. A contrast enhanced MRI showed an extra-axial mass in the left parietal region, with associated effacement of adjacent sulci, which demonstrated heterogeneous enhancement following contrast administration with associated effacement of adjacent sulci. It measured 3.9 × 1.1 cm in maximum transverse dimensions and extended supero-inferiorly for approximately 3.5 cm. There was also an enhancing dural tail, and the pattern was found to be most consistent with meningioma. She was discharged with pain medications and shortly thereafter was seen in neurosurgery clinic.

Because of her persistent headaches and pain on the left side of the head when she lay down, she

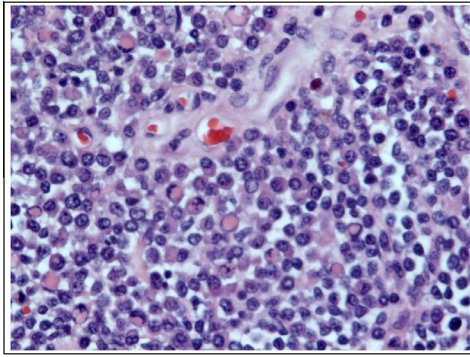


Fig. 1. Lymphoplasmacytic lymphoma showing intranuclear inclusions.

underwent a resection of the left parietal mass. Pathology showed that the mass was generally composed of small lymphoid cells with scattered plasmacytoid cells. Mature plasma cells were also noted. Intranuclear inclusions (Russell bodies) were also seen in some of the plasmacytoid cells (Fig. 1). This pattern of lymphocytes and plasmacytoid cells with Russell bodies was consistent with lymphoplasmacytic lymphoma. Flow cytometry demonstrated monoclonal B cells that were negative for CD5 and CD10, also consistent with this diagnosis. Her serum immunoglobulin levels were IgG 1220 mg/dL, IgM 317 mg/dL and IgA 130 mg/dL. Her total protein was 6.2 g/dL. Serum protein electrophoresis did not show any distinct paraprotein but immunofixation showed abnormal monoclonal IgM and lambda bands. Her beta microglobulin level was 1 mg/L, LDH 131 IU/dL. Her hepatitis panel was positive for hepatitis C antibody. A lumbar puncture was done and cerebrospinal fluid showed a total WBC of 2/ μ L with 4% lymphocytes; cytology was negative for malignant cells and no monoclonal bands were seen on immunofixation. Three weeks after this diagnosis she was admitted with shortness of breath, was found to have a pulmonary embolism, and was started on enoxaparin. A PET CT scan was done to look for other sites of lymphoma but this was negative with no FDG avid lesions to show active lymphoma. A bone marrow biopsy was performed which showed no lymphoma involvement. Bone marrow was negative by flow cytometry and cytogenetics showed normal karyotype. Because of the localized nature of the lymphoma and absence of either malignant cells or monoclonal protein in the CSF, we decided to treat with radiotherapy alone. She was treated with a total dose of 5040 cGy in 40 fractions

to the tumor bed. After 1 year of follow up, there is no evidence of disease recurrence.

DISCUSSION

Lymphoplasmacytic lymphoma (LPL, previously termed lymphoplasmacytoid lymphoma) is an uncommon mature B-cell lymphoma usually involving the bone marrow and, less commonly, the spleen and/or lymph nodes.²

PCNSL is defined as non-Hodgkin lymphoma (NHL) arising within the CNS and confined to it at the time of diagnosis.³ Interestingly, the incidence of PCNSL has been increasing at a rate higher than peripheral non-Hodgkin lymphomas (NHL) or glial tumors.⁴ PCNSL is characterized by a supratentorial (and very rarely spinal) localization, intensive and homogeneous contrast enhancement, minor or moderate edema, absence of necrosis, and proximity to the ventricles.⁵ Ninety-five percent of PCNSLs are diffuse large B-cell lymphomas (DLBCL).⁶ Other lymphoma categories like Burkitt's, lymphoblastic, marginal zone, and small lymphocytic lymphoma are uncommon differential diagnoses. T-cell PCNSLs are ~2% of cases in Western countries.⁷

The largest case series of CNS low grade lymphomas reported 40 cases, which include eight T-cell lymphomas and 32 B-cell lymphomas. The low grade B-cell lymphomas described in this series were predominantly non-classifiable small lymphomas but 11 cases of LPL and one case of follicular lymphoma were noted.⁸ In another study of 15 patients with low grade primary CNS lymphoma, only two were found to have lymphoplasmacytic lymphoma.⁹ A study by Bogdahn et al in 1986 of 10 patients with primary CNS low grade lymphomas had four patients with lymphoplasmacytoid immunocytoma.¹⁰ Besides these case series, we found four other cases of CNS lymphoplasmacytic lymphoma in the literature.^{11–14} The confirmation of the diagnosis and further specification may be difficult in PCNSL due to the small amount of biopsy material, which, however, is a general problem in brain tumors diagnosed by stereotactic biopsy.

The major diagnostic dilemma we had was to differentiate primary CNS lymphoplasmacytic lymphoma from Bing-Neel syndrome. In a review by Fintelmann et al.¹⁵ the authors proposed to limit the definition of Bing-Neel Syndrome to patients with proven Waldenström's macroglobulinemia and CNS symptoms not due to hyperviscosity or large cell transformation. But for diagnosis of Waldenström's macroglobulinemia, bone marrow involvement is a major criterion. Because bone marrow biopsy was

negative in our patient, a diagnosis of primary CNS lymphoplasmacytic lymphoma was made.

As low grade primary CNS lymphomas are very rare, there are no set guidelines for treatment. Carrasco et al reported a pituitary LPL patient who maintained complete remission for 4 years after transsphenoidal surgery, chemotherapy and radiotherapy.¹⁴ Braks et al. (2000) reported a 42-year-old woman with a 3-year history of headaches and vertigo and a 1-year history of seizures. The tumor, histologically an immunocytoma, remained unchanged throughout the entire observation period with anticonvulsive treatment alone.¹¹ Another study reported one patient with a spinal cord LPL at T4 level. The patient received only surgical resection (corpectomy) without neurologic se-

quela and has lived without relapse for 51 months.⁹ In the case of our patient, we decided to treat her only with radiation.

We present here a very rare case of primary CNS lymphoplasmacytic lymphoma. So far in the literature only 21 cases have been reported. This case emphasizes that differentiating between a low grade CNS B-cell lymphoma from a DLBCL or a reactive phenomenon is essential, both from a therapeutic and a prognostic standpoint.

CONFLICT OF INTEREST

None declared.

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